Acute Aortic Syndrome (AAS)

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Overview

• Acute aortic syndrome background
• Review aortic anatomy
• Menu of tests
• Aortic dissection
  – Index patient AD
  – Discussion
• Intramural hematoma
  – Companion patient 1
  – Discussion
• Ruptured aneurysm
  – Companion patient 2
  – Discussion
AAS Facts

• Spectrum of *life-threatening* aortic emergencies involving non-traumatic acute aortic injury
  – Aortic dissection
  – Intramural hematoma
  – Penetrating atherosclerotic ulcer
  – Thoracic aortic aneurysm rupture
AAS Epidemiology and Risk Factors

• Epidemiology
  – 2.6-3.5 cases per 100,000
  – 2/3 male
  – Average age 63 years

• Risk factors
  – Chronic hypertension
  – Atherosclerosis
  – Weakening of aortic media
  – Vasa vasorum with predisposition to rupture

Shiau et al, 2010
AAS Symptoms

• Symptoms
  – Chest pain
    • Severe, tearing, migratory
  – Neck, throat pain
    • May indicate injury to ascending aorta
  – Back, abdominal pain
    • May indicate injury to descending aorta
  – Syncope
    • Complications secondary to AAS (hypoperfusion)
Review of Aortic Anatomy

Adapted from clevelandclinic.org
http://my.clevelandclinic.org/heart/disorders/aorta_marfan/aortaillust2.aspx
Dissection vs Intramural Hematoma

From Shiau et al, 2010

IMH=intramural hematoma
TL=true lumen
FL=false lumen
## Imaging studies for acute aortic syndrome

<table>
<thead>
<tr>
<th>IMAGING STUDY</th>
<th>ADVANTAGES</th>
<th>DISADVANTAGES</th>
</tr>
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</table>
| Cardiac-gated multidetector computed tomography | Highly specific and sensitive  
Can diagnose major causes of acute aortic syndrome  
Rapid scan and interpretation times | Large doses of ionizing radiation and contrast                                |
| Chest radiography                          | Very rapid result  
Very helpful to exclude nonaortic causes | Low-to-moderate specificity for acute aortic syndrome  
Low sensitivity for aortic pathology |
| Transesophageal echocardiography            | Highly specific and sensitive for ascending aortic dissection and aneurysmal disease | Requires skilled personnel to perform and interpret  
Often unavailable in the emergency department |
| Angiography                                | Highly sensitive and specific for aortic dissection and aneurysmal disease | Invasive  
Requires contrast  
Cannot diagnose intramural hematoma |
| Magnetic resonance imaging                 | Highly specific and sensitive  
Can diagnose major causes of acute aortic syndrome  
Can be accurate without using contrast | Difficult to arrange in an emergency  
Prolonged scanning time and limited ability to manage unstable patients during scan |

From Smith and Schoenhagen, 2008
# ACR Appropriateness Criteria for Aortic Dissection

## Clinical Condition:
**Acute Chest Pain — Suspected Aortic Dissection**

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
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</thead>
<tbody>
<tr>
<td>X-ray chest</td>
<td>9</td>
<td>Should be performed if readily available at the bedside and does not cause delay in obtaining a CT or MRI. Alternative causes of chest pain may be discovered. Not the definitive test for aortic dissection.</td>
<td>0</td>
</tr>
<tr>
<td>CTA chest and abdomen</td>
<td>9</td>
<td>Recommended as the definitive test in most patients with suspicion of aortic dissection.</td>
<td>💫</td>
</tr>
<tr>
<td>MRA chest and abdomen with or without contrast</td>
<td>8</td>
<td>Alternative to CTA for: contraindication to CT (iodinated contrast), multiple prior chest CTA for similar symptoms, and in patients showing no signs of hemodynamic instability. Scanner availability and local expertise limit widespread use as there is potential for delay in diagnosis. See statement regarding contrast in text under “Anticipated Exceptions.”</td>
<td>O</td>
</tr>
<tr>
<td>US echocardiography transesophageal</td>
<td>8</td>
<td>If skilled operator readily available.</td>
<td>O</td>
</tr>
<tr>
<td>Aortography thoracic</td>
<td>5</td>
<td></td>
<td>💫</td>
</tr>
<tr>
<td>US echocardiography transthoracic</td>
<td>4</td>
<td></td>
<td>O</td>
</tr>
</tbody>
</table>

**Rating Scale:** 1, 2, 3 Usually not appropriate; 4, 5, 6 May be appropriate; 7, 8, 9 Usually appropriate

*Relative Radiation Level*
Patient AD: Clinical History

• Chief Complaint: 75 year old female with chest pain, abdominal pain, back pain, and nausea/vomiting

• Past Medical History:
  – Hypertension
  – End-stage renal disease on peritoneal dialysis six years s/p transplant
Our patient did not have a chest x-ray (CXR) performed at the time of her presentation, probably due to a triple rule-out (pulmonary embolism, acute coronary syndrome, and acute aortic syndrome) protocol best done on CT.

Let’s take a look at a baseline CXR done 2 years ago, and then another CXR after dissection repair done for concern of another dissection.
Patient AD: Comparison of CXR

r/o dissection (from later timepoint)

Widened mediastinum

baseline

PACS, BIDMC
Patient AD: Dissection at aortic root on CT

C+ Axial CT

- Intimomedial flap in ascending aorta
- Mediastinal hematoma
- False lumen: hypodense compared to true lumen
- True lumen: hyperdense compared to false lumen
- Contrast extravasation into anterior mediastinum
- Intimomedial flap in aortic arch
Patient AD: Aortic dissection through subclavian and common carotid

C+ Axial CT

Intimomedial flap in right subclavian

Intimomedial flap in right common carotid

Intimomedial flap in left common carotid
Patient AD: True lumen supplying celiac trunk on CT

C+ Axial CT

Celiac trunk
Patient AD: Dissection involves the SMA

C+ Axial CT

Intimomedial flap in SMA

Takeoff of the SMA
Patient AD: True lumen supplies renal arteries

C+ Axial CT

Takeoff into renal arteries
Patient AD: Aortic dissection inferior to level of renal arteries on CT

C+ Axial CT
Patient AD: Dissection affects ascending arch

C+ Sagittal CT
Patient AD: Descending arch dissection on CT

Anterior true lumen supplies celiac artery and SMA

C+ Sagittal CT
PACS, BIDMC
Patient AD: Follow-up

• AD was taken emergently to OR, and had replacement of her ascending aortic dissection with Dacron graft.
• She is alive and well today.
Aortic Dissection Facts

• Most common cause of AAS
• 38% mortality by 24 hours and 50% mortality by 48 hours if undiagnosed and untreated
• Etiology
  – Systemic hypertension
  – Intramural hematoma
• Pathophysiology
  1. Laceration in aortic intima
  2. Pressurized blood enters into and dissects between layers of aortic media, creating false lumen
Aortic Dissection Diagnosis

• Diagnosis
  – CXR
    • Widened mediastinum
    • Widened aortic knob
    • Calcium sign
    • Pleural effusions
  – Contrast-enhanced CT angiography
    • Visualize true and false lumens, intimomedial flap, and often intimal tear
    • True lumen has increased contrast density
    • Pulsation artifact can be corrected with ECG gating
    • Coarctation, pseudocoarctation, saccular aneurysm, and pseudoaneurysm can mimic dissection on CTA
Aortic Dissection Classification and Complications

• Classification
  – Type A: ascending aorta
    • Requires immediate surgical attention
  – Type B: dissection in descending aorta distal to left subclavian artery origin

• Complications
  – Aneurysm formation
  – Rupture
  – Continuation of dissection into pericardium +/- tamponade
  – Extension into carotid or coronary arteries causing stroke or MI, respectively
  – Renal and mesenteric ischemia secondary to hypoperfusion due to dissection extension into branch vessels or branch-vessel perfusion
  – Rare complication: rupture into mediastinum
Companion Patient 1: Intramural hematoma on C-CT

C- Axial CT

Hematoma

Aortic lumen

Hematoma
Companion Patient 1: Intramural hematoma on C+ CT

C+ Axial CT

No contrast in intramural hematoma

Contrast-filled aorta

Possible extravasated contrast
Intramural Hematoma Facts

• Hemorrhage within media
  – **Absence** of intimomedial tear

• Etiology
  – Focal penetrating ulceration in atherosclerotic plaque
  – Thrombosed dissected lumen

• Pathophysiology
  – Hypertension injures media→spontaneous rupture of vasa vasorum and mechanical strain→intimal tear
IMH Diagnosis

• Diagnosis
  – Not differentiated clinically from dissection
  – Imaging
    • Pre-contrast imaging needed to evaluate possible IMH
    • Acute IMH: relatively circumferential crescent or ring-like hyperdensity along margin of aorta
      – Looks like “thickened aortic wall”
      – There will be no contrast enhancement of this area
    • Subacute or resolving IMH
      – Isodense to blood on pre-contrast CT
IMH Differential and Treatment

• Differential Diagnosis
  – Aortic-wall thickening such as aortitis
  – Mural thrombus
  – Soft atheromatous plaque

• Treatment
  – 10% will regress spontaneously
  – 20-45% will progress to pseudoaneurysm
  – 2-20% become acute aortic dissections
  – Requires close monitoring
Companion Patient 2: Ruptured Aneurysm on C-CT

Hemothorax

69.61mm
Companion Patient 2: Ruptured Aneurysm on C+ CT

Extravasated contrast

Contrast-filled aneurysm

PACS, BIDMC
Ruptured Aneurysms Facts

• Aortic aneurysm may enlarge rapidly, show signs of imminent rupture, or have already ruptured

• Aortic thoracic aneurysm
  – Permanent localized dilatation >50% of normal, or >5cm and involving all 3 layers of aortic wall

• Symptoms:
  – Chest or back pain, but patient not clinically hypertensive (although has PMH of HTN)
Ruptured Aneurysms on CT Imaging

• CT signs of imminent rupture
  – IMH
  – Discontinuous calcification in circumferentially calcified aorta “missing calcium sign”

• CT signs of rupture
  – Mediastinal hematoma
  – Stranding of periaortic fat
  – Hemothorax
  – More rarely: pericardial effusion
Laplace’s Law

• Laplace’s law: \( T = P \times R \)
• \( T \)= wall tension
• \( P \)= pressure (increased in HTN)
• \( R \)= radius (increased in aneurysm)
Summary 1

• Acute aortic syndrome is a spectrum of disorders of non-traumatic injury to the aorta
  – Aortic dissection

• We saw an example of this involving both the ascending and descending portions of the aorta with involvement through the subclavian arteries and past the level of the renal arteries
• Stanford Type A involves the ascending portion
• Stanford Type B involves the descending portion
Summary 2

– Intramural hematoma
  • We saw an example of an intramural hematoma that was seen both on contrast negative and contrast-enhanced CT
  • Can cause both ruptured aneurysm and dissection

– Ruptured aneurysm
  • We saw an example of ruptured aneurysm causing hemothorax
  • Laplace’s law explains why an aneurysm can rupture
References

• Aorta Illustration 2. Cleveland Clinic. http://my.clevelandclinic.org/heart/disorders/aorta_marfan/aortaillust2.aspx. 1/22/01
• Villacosta I, Roman JA. Acute aortic syndrome. Heart. 2001;85:365-8
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